ASD was a risk factor for major complications (p=0,03). The correlation coefficient between the size of the ASD with TEE and ICE was 0.887.

Conclusion: In an unselected patient population, ICE provides a safe and efficient guidance for device closure of ASD, even for large defects with deficient rims.

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Long-term outcome of 117 patients with univentricular heart and common atrioventricular valve

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Introduction: Few studies investigated the long-term outcome of patients with univentricular heart and common atrioventricular valve.

Method: We retrospectively analysed the medical files of all patients univentricular heart with common atrioventricular valve in the setting of heterotaxy or with unbalanced atrioventricular septal defect preventing biventricular repair.

Results: 117 patients were identified during the study period. 89/117 had a postnatal diagnosis. 28/117 patients never underwent surgery, 25/117 underwent one palliation surgery (Blalock-Taussig-shunt (BTS)/ pulmonary banding), and finally, 61/117 patients entered a sequential cavopulmonary connection program: 37/61 had partial cavopulmonary connection at the time of data analysis while 24/61 had total cavopulmonary connection (TCPC). The average age at TCPC was 7.6 years +/-4 years [1.7-16 years]. Three patients were eventually transplanted.

The overall mortality was 59% (69/117): 65% and 30% in heterotaxy and in patients with unbalanced atrioventricular septal defect respectively. Mortality was 85% (24/28) in the subgroup of patients who never underwent surgery, 93% in the subgroup of patients who had a BTS, and 89% after pulmonary banding. In the subgroup planed to have TCPC, 49% died after partial cavopulmonary connection and survival rate was 71.6% [50.7-100] in patients who had TCPC.

Conclusion: The long-term outcome of univentricular hearts with common atrioventricular valve treated in a tertiary referral center showed a high mortality rate. Patients with this kind of complex congenital heart disease should undergo the sequential univentricular program without delay in order to obtain better long-term survival.

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Toward better ventricular pacing in patients with a systemic right ventricle

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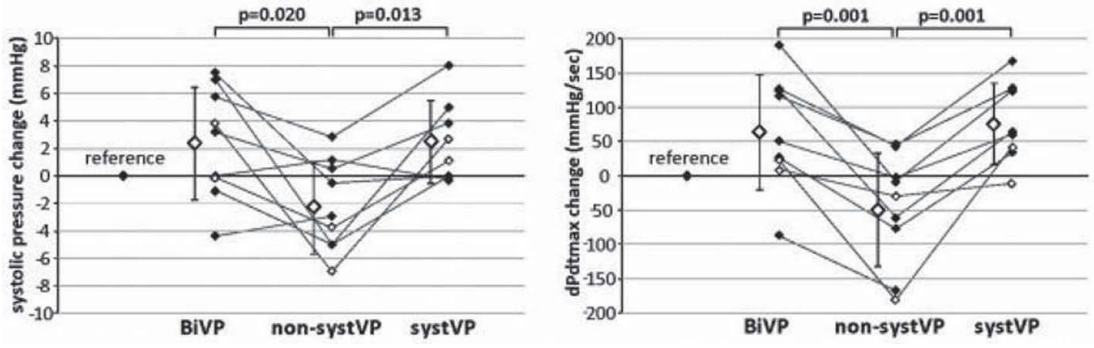
Introduction: Patients with transposition of the great arteries (TGA) and atrial redirection, have an important risk of heart failure caused by dysfunction of the systemic RV. Conventional non-systemic ventricular pacing (non-systVP) may even further increase this risk. We investigated whether these patients may benefit from biventricular pacing (BiVP) and/or single-site systemic ventricular pacing (systVP).

Methods: During clinically indicated catheterization in 9 patients with TGA and status post Senning procedure, endocardial ventricular stimulation (overdrive DDD, 80-90 bpm) was applied with temporary pacing leads at the non-systemic ventricle and the systemic ventricle. Acute changes in dP/dtmax and systolic pressure of the systemic ventricle, as induced by non-systVP, systVP and BiVP compared to reference, were assessed with a pressure wire (RADI Medical Systems®) within the systemic ventricle. Reference was AAI pacing with similar heart rate (n=7; filled squares), or non-systVP at a lower heart rate than during stimulation at experimental sites (85 vs. 90 bpm; n=2).

Results (Figure): Systemic dP/dtmax and systolic ventricular pressure were significantly higher during systVP (+15.6% and +5.1%) and BiVP (+14.3% and +4.9%) when compared with non-systVP. In 6 out of 7 patients, dP/dtmax was even higher during BiVP and systVP than during AAI pacing.

Conclusions: Patients with systemic RV, such as patients with TGA and atrial redirection, may benefit from biventricular or systemic ventricular pacing, especially when ventricular pacing is indicated.



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Is valvulation of Fontan circulation of clinical value?

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Objectives: Fontan circulations conduct to complications such as heart dysfunction, arrhythmia, exsudative entheropathy. The circulation largely depends on breathing and gravity. A part of the caval flow comes back in the abdominal compartment. We propose to study the insertion of valve in the system in patients with refractory dysfunction. We hypothesized that the valve will reduce the effects of the respiration and gravity improving the net flow in the lungs, the exercise capacity and the enteropathy. The Melody valve is the valve of choice because it is fine and harvested from the bovine jugular vein.

Methods: All patients who received a valve in a Fontan circulation were eligible.

Results: four patients were included. All had a Fontan circulation with clinical and hemodynamic alterations: 2 patients had refractory exsudative

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entheropathy, one a dyfunction of the systemic ventricle and edema, and the last one severe venous insufficiency of the legs. A Melody valve was successfully inserted in all patients; three in the pathway, one after the iliac bifurcation. There was no significant modification of the pressures after valvulation. No acute complication was recorded. There was an improvement of the doppler through the proclive and breathing. However, these were not associated with significant clinical improvement. No thrombosis of the valve accured during the short follow-up (1 to 6 months).

Conclusions: the valvulation of the Fontan circuit is easily performed. We noticed an improvement of the hemodynamic and flow. However, no clinical improvement was recorded as a result. A longer follow-up is needed to appreciate the risks as well as the interest of this procedure.

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Percutaneous insertion of a Melody valve in tricuspid position: technical aspects

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Background: percutaneous transcatheter heart valve replacement of aortic or pulmonary valve is established. Transcatheter atrioventricular valve replacement is been described. We report our experience focusing on the technical aspects.

Methods: we retrospectively review the files of patients who received a transcatheter valve in tricuspid position between 2008 and 2012.

Results: Four patients were found. 3 had a heterograft (conduit of 14-mm, Sorin 33 et Edwards Perimount 33) and one had a connection between the RA and the RV infundibulum without a valve. Two patients had tricuspid regurgitation as a primary lesion, one had stenotic valve and the last one a mixted lesion. All successfully received a Melody valve from a femoral access. In patients with stenotic lesion, a predilatation using a high pressure balloon was performed before valve implant. In patients with regurgitation, the landing zone was calibrated using a low pressure balloon. These patients were presented to create a landing zone of adequate diameter. Melody valves were inserted using a 22-mm balloon catheter in 3 and a 24-mm in one. All but one were post-dilated. There was no significant regurgitation. The mean gradient across the tricuspid valve felt from 12 to 4.6-mmHg. One patient needed an epicardic pacemaker because of AV block following balloon dilatation. One patient required inotropic support and ventilation following the procedure but recovered after few days.

Conclusion: Transcatheter tricuspid valve insertion is feasible in patients with surgical hetero or homografts after a careful selection. The mechanism of dysfunction must be known. In case of stenosis or mixted lesions, the only question is to know if the stenosis could be relief. In case of regurgitation, it is very important to know the features of surgical substrats and to calibrate the tricuspid orifice. Finally, patients with inappropriate landing zone should be presented prior to valve insertion.

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Potts' shunt in children with idiopathic pulmonary arterial hypertension: long-term results

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Background: Idiopathic pulmonary arterial hypertension (IPAH) remains a progressive fatal disease. Palliative Potts'shunt has been proposed in children displaying supra-systemic IPAH.

Methods: A retrospective multicenter study to evaluate Potts'shunt in pediatric IPAH.

Results: Between 2003 and 2010, 8 children with supra-systemic IPAH and in WHO functional class IV despite medical PAH therapy underwent Potts'shunt. Age at IPAH diagnosis ranged from 4 to 180 months (median age: 64 months). Surgical procedure was performed in a mean delay of 41.9±54.3 months (from 4 to 167 months, median delay: 20 months) after IPAH diagnosis. Mean size of the Potts'shunt was 9.25±3.30 mm. Two patients, whose medical PAH therapy had been interrupted just after surgery, died at post-operative day 11 and 13 from acute pulmonary hypertensive crisis. After a mean follow-up of 63.7±16.1 months, the 6 children who were discharged from hospital were alive. Functional status improved markedly in the 6 survivors with a WHO functional class I (n=4) or II (n=2) at last follow-up, consistent with significant improvement of 6 minute-walk distance [302±95 (51±20% of theoretical values) vs 456±91 meters (68±10% of theoretical values), p=0.038] and decrease of brain natriuretic peptid (BNP) levels (608±109 vs 76±45 pg/ml, p=0.035). No Potts'shunt was found restrictive at last echocardiography.

Conclusion: Palliative Potts'shunt constitutes a new alternative to lung transplantation in severely ill children with supra-systemic IPAH, carrying a prolonged survival and persistent improvement in functional capacities.

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Conotruncal defects: is the ventricular septal defect always the same?

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Conotruncal defects (CTD) are a group of cardiac malformations heterogeneous from an anatomic standpoint but with a common embryologic origin: an abnormal rotation of the outflow tract. The outlet septum is therefore malaligned or absent, resulting in a ventricular septal defect (VSD).

Aim of the study: To analyze the anatomy of the VSD in hearts with CTD.

Material and methods: We reviewed 200 heart specimens with CTD from the anatomic collection of the French Center of Reference for Complex Congenital Heart Defects: 70 Tetralogy of Fallot (TOF), 53 TOF with pulmonary atresia (TOF-PA), 54 common arterial trunk (CAT), and 23 interrupted aortic arch type B (IAA-B). Special attention was paid to the rims of the VSD viewed from the right ventricular side, the relationships between tricuspid and aortic valves, and the anatomy of the outlet septum.

Results: The VSD was located between the 2 limbs of the septal band (conoventricular) in all hearts. There was a fibrous continuity between tricuspid and aortic valves in 0% of IAA-B, 66% of TOF, 37% of TOF-PA, 1% of CAT ($p<0.005$). When present, this continuity always involved the anterior tricuspid leaflet. The outlet septum was demonstrable in 81% of IAA-B, 96% of TOF, 39% of TOF-PA, 0% of CAT ($p<0.0001$).

Conclusion: All CTD share the same VSD, located between the two limbs of the septal band. However, there are some differences regarding the inferior rim of the VSD. The continuity of the aortic valve with the anterior, and not the septal, tricuspid leaflet indicates that this continuity may be a consequence of the malposition of the ventriculo-infundibular fold, along with its outlet septal component, rather than a perimembranous extension of the VSD. Finally, these differences suggest an anatomic continuum from IAA-B to CAT rather than distinct physiological phenotypes, related to various degrees of abnormal rotation of the outflow tract, excessive in IAA-B, incomplete in TOF, TOF-PA and CAT.

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Congenital left coronary ostial atresia or stenosis – a series of four neonatal fatal cases

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